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NASAL MANIFESTATIONS OF GRANULOMATOSIS WITH POLYANGIITIS: A CLINICAL REVIEW

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ABSTRACT

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare autoimmune vasculitis characterized by granulomatous inflammation and necrotizing vasculitis of small- to medium-sized vessels. The nasal cavity is one of the most frequently involved sites and often presents as the first manifestation of disease. This review explores the clinical features, diagnostic methods, and therapeutic approaches for nasal involvement in GPA. A literature review was performed to synthesize current understanding of the disease. Findings suggest that nasal crusting, epistaxis, septal perforation, and saddle nose deformity are among the most common symptoms. Diagnosis relies on clinical suspicion, positive ANCA serology, and confirmatory histopathology. Immunosuppressive therapy remains the cornerstone of treatment, with surgical interventions reserved for structural complications. Prompt diagnosis and a multidisciplinary approach can improve patient outcomes.

KEYWORDS: Granulomatosis with polyangiitis; vasculitis; saddle nose deformity; immunosuppressive therapy; nasal biopsy; necrotizing granuloma.

INTRODUCTION

Granulomatosis with polyangiitis (GPA) is a rare systemic vasculitis affecting primarily the upper and lower respiratory tracts and kidneys. Previously referred to as Wegener's granulomatosis, it is now recognized under its current name to better reflect the underlying pathology and to move away from historical eponyms. Nasal and sinus involvement is observed in over 85% of cases, often preceding systemic symptoms such as renal or pulmonary involvement.[1] Common symptoms include nasal obstruction, chronic rhinorrhea, crusting, epistaxis, and septal perforation. Structural deformities such as saddle nose may occur in longstanding, untreated cases. This review focuses on the pathogenesis, clinical presentation, diagnosis, and management of GPA with a special emphasis on nasal manifestations.

MATERIALS AND METHODS

A structured literature review was conducted using PubMed, Google Scholar, and Scopus databases for studies published between 2000 and 2024. Search terms included "Granulomatosis with polyangiitis," "Wegener's granulomatosis," "nasal vasculitis," "ANCA-associated vasculitis," and "saddle nose deformity." Articles were selected based on relevance to nasal and sinonasal involvement in GPA, including original research, clinical reviews, and case reports. References from key articles were manually reviewed for additional sources. Only English-language publications were considered.

RESULTS

Nasal involvement in GPA is frequently the earliest sign and may mimic chronic rhinosinusitis.[2,3] Patients often report symptoms such as nasal crusting, purulent discharge, epistaxis, and nasal obstruction. On examination, septal perforation and mucosal ulceration may be seen, and in severe or chronic cases, collapse of the nasal bridge—saddle nose deformity—may develop due to cartilage destruction.[4]

Diagnostic evaluation includes nasal endoscopy and biopsy, which may reveal granulomatous inflammation with necrosis and vasculitis.[5] Imaging such as CT scans of the sinuses often shows mucosal thickening, bone erosion, or sinus opacification.[6] Serologic testing is critical; cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA), primarily directed against proteinase-3, are present in 80–90% of active systemic cases.[7] However, ANCA may be negative in localized disease.[8]

Treatment involves induction of remission with immunosuppressive agents such as corticosteroids in combination with cyclophosphamide or rituximab.[9] Methotrexate may be used in milder forms. Maintenance therapy includes azathioprine, methotrexate, or mycophenolate mofetil.[10] Surgery may be indicated for septal repair or saddle nose correction once disease is controlled.[11]

DISCUSSION

Nasal manifestations of GPA are significant both for early detection and monitoring of disease activity. Crusting and bleeding are among the most common complaints, and saddle nose deformity is a hallmark of advanced disease.[4] Histologically, the triad of necrosis, granulomatous inflammation, and vasculitis is diagnostic, though biopsy findings may be non-specific, especially in early disease.[5]

ANCA serology, though not always present in limited disease, is a useful diagnostic marker and helps differentiate GPA from other causes of chronic rhinosinusitis.[7] Immunosuppressive therapy has revolutionized the prognosis of GPA, reduced mortality and preserving organ function when initiated early.[9] However, relapses are common and require long-term monitoring.[10]

Multidisciplinary management, including rheumatologists, ENT specialists, and pathologists, is essential for optimal patient outcomes. In cases with disfiguring nasal damage, reconstructive surgery can significantly improve quality of life.[11]

CONCLUSION

Granulomatosis with polyangiitis frequently presents with nasal and sinonasal symptoms that can mimic benign ENT conditions. Recognizing these signs early, supported by histopathology and ANCA serology, is essential for timely diagnosis. Immunosuppressive treatment is effective in controlling disease activity and preventing complications. A team-based, individualized approach to therapy and monitoring is essential for managing this complex condition.

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